Diagnostic Dilemma: Guillain Barre syndrome with brisk reflexes

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GBS is an acute monophasic immune mediated polyradiculoneuropathy which essentially presents with progressive motor weakness and areflexia with variable sensory disturbances.1 Two distinctive pathological subtypes of GBS have been mentioned –axonal and demyelinating. In Chinese, Japanese and European population, there have many case reports mentioned of another possible subtype of GBS, i.e., GBS with preserved or exaggerated reflexes.1,2 This entity is rare in Indian subcontinent and only a few cases have been reported.3,4 Unless a high index of suspicion is maintained, such case is often missed.

A 14 year old boy with no significant past medical history presented to us with the history that 7 days back he experienced tingling sensation in both hand followed by difficulty in handling mobile phone buttons. Next day he noticed difficulty in walking followed by difficulty in climbing stairs which progressed over next 2-3 days so that he was not able to walk without support of 2 persons and also was not able to sit up from supine position. On 4th day, he noticed difficulty lifting both arms above head followed next day by mild difficulty in lifting head from pillow. However, he never experienced any difficulty in breathing, swallowing, closing eyes or any bladder symptoms. There was no history any antecedent illness or recent vaccination or toxin exposure. On examination, flaccidity was noted in all group of muscles. Power in proximal and distal lower limbs was grade 2 (MRC grading) and in proximal and distal upper limbs was grad 4- and grade 3 respectively. Truncal and neck weakness was also noted. Deep tendon reflexes were brisk throughout the course of illness and superficial reflexes (plantar and abdominal) were normal. Sensory and cerebellar examination was normal. Investigations revealed normal creatine kinase and normal potassium level. MRI brain with cervical spine was normal. Nerve conduction study showed pure motor axonal polyneuropathy with absent F waves suggesting axonal variant of GBS. Albumino-cytological dissociation was noted on CSF examination (cells-3, proteins-83, normal sugar). Electromyography was done which showed decreased recruitment of motor unit potentials with normal morphology of MUAPs and without any spontaneous activity. Patients illness was static for 3 days and since the time he presented to us. He was managed conservatively and he started improving over next 3 weeks. On discharge, his power was grade 4- in lower limbs and 4+ in upper limbs and was able to walk with support on his own. Repeat nerve conduction studies after 4 weeks showed improvement in CMAP amplitudes to almost normal with imperisent F wave.

GBS with normal or brisk reflexes is uncommon in India. The variants which have described with normal or brisk reflexes are AMAN, acute motor conduction block neuropathy and acute facial diplegia with brisk reflexes.1,5,6 A rapid recovery has been noted in patients with preserved reflexes, preceding Campylobacter jejuni or H. influenzae infection and positive anti-GM-1 ganglioside antibodies.12 Antibody testing is not freely available in India which makes the diagnosis, analysis and further correlation more difficult.

The proposed mechanism for brisk deep tendon reflexes is dysfunction of inhibitory systems in spinal interneurons.6 The pathophysiological mechanism is supposed to be due to distal conduction disturbances rather than axonal degeneration which produces low amplitudes of motor unit potentials on nerve conduction studies. This is termed as reversible conduction failure or acute motor conduction block neuropathy.7 The presumed reason is conduction disturbances at node of Ranvier rather than demyelination. This motivated us to go for a needle EMG which showed decreased recruitment suggestive of early neurogenic pattern. EMG has not been done in earlier cases reported from India.3 High cervical myelopathy and hypokalemia are the most important differential diagnoses of GBS with normal or brisk reflexes. However, a careful history, detail physical examination and thorough investigations help to avoid confounders and achieve correct diagnosis.

References